

## THE MODIFIED FONTAN OPERATION

### An analysis of risk factors for early postoperative death or takedown in 702 consecutive patients from one institution

To better understand risk factors associated with early postoperative death or failure, we reviewed our entire experience with 702 consecutive patients who had the modified Fontan operation at the Mayo Clinic between October 1973 and December 1989. The event rate for takedown of repair or death during the initial hospitalization or within 30 days of the operation was 14.8% (successful takedown of the repair,  $n = 6$ ; death,  $n = 98$ ). To identify variables associated with early death or Fontan takedown, we analyzed 33 clinical and hemodynamic variables in a univariate and multivariate manner. On the basis of a stepwise logistic discriminant analysis, patients who were younger and operated on before 1980 with a higher preoperative pulmonary artery mean pressure, asplenia, higher intraoperative (after Fontan operation) right atrial pressure, longer aortic crossclamp time, and pulmonary artery ligation were more likely to have the outcome event of interest ( $p$  values  $<0.05$ ). A new variable, corrected pulmonary artery pressure (that is, mean preoperative pulmonary artery pressure divided by the ratio of pulmonary to systemic flow if the ratio of pulmonary to systemic flow is greater than 1.0), was significantly associated with the outcome event univariately ( $p = 0.002$ ), but was no more predictive than the preoperative pulmonary artery mean pressure. Variables less predictive of the outcome event in this analysis included multiple prior operations, polysplenia syndrome, complex anatomy other than asplenia syndrome, and systemic atrioventricular valve regurgitation. These results represent the largest single-institution review of the Fontan operation and suggest that some anatomic and hemodynamic variables previously predictive of poor early outcome have been nullified by current operative methods. (*J THORAC CARDIOVASC SURG* 1995;109:1237-43)

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Since the first successful Fontan operation for tricuspid atresia was reported in 1971,<sup>1</sup> numerous modifications have been proposed as definitive palliation for an increasing variety of complex cyanotic congenital heart defects with a functional single ventricle.<sup>2-19</sup> More recently, surgical modifications have focused on staging the separation of systemic and pulmonary circulations<sup>9-13</sup> in an attempt to reduce the early mortality and morbidity

associated with the modified Fontan operation in patients with higher risk factors.

As experience was gained with the modified Fontan operation at the Mayo Clinic and more patients with complex cyanotic heart disease requiring a one-ventricle repair were referred, our selection criteria were broadened to accommodate these high-risk patients, often with encouraging early results.<sup>20-24</sup> However, apart from modifications for specific technical difficulties,<sup>3, 4, 6, 25</sup> the operation has remained essentially unchanged throughout the years, allowing this analysis of risk factors for early failure to be undertaken with little influence of surgical variation.

#### Patients and methods

**Study group.** Since the first operation at the Mayo Clinic in October 1973 and as of the end of 1989, 702 consecutive patients had the modified Fontan operation. These 702 patients form the basis of this review. All but 8 of these 702 patients known to be alive at the date of the most recent follow-up had more than 30 days of postoperative follow-up.

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The age at operation ranged from 8 months to 42 years, and the median age was 9 years. Forty percent of the patients were female. The primary cardiac defect was tricuspid atresia in 203 (28.9%), double-inlet ventricle in 226 (32.2%), and complex cardiac defects in 273 (38.9%).

**Surgical technique.** The surgical techniques have been described fully elsewhere<sup>15, 18, 21</sup> and may be summarized as follows: the operation usually was done with the use of deep hypothermia (20° to 25° C) facilitated by periods of low-flow perfusion (0.5 to 1.5 L/min per square meter) or circulatory arrest as necessary. Intermittent cold blood or crystalloid cardioplegia and topical hypothermia were used for myocardial protection. Direct atriopulmonary connection was the preferred anastomosis for tricuspid atresia and double-inlet left ventricle. In most patients with univentricular heart, the systemic and pulmonary venous drainage was partitioned in such a way that the coronary sinus drained into the pulmonary venous side of the atrial baffle, but no special techniques were used to achieve this if the anatomy was unfavorable. Partial Fontan techniques such as the fenestrated Fontan procedure,<sup>12, 13, 26</sup> adjustable atrial septal defect Fontan procedure,<sup>10, 11, 27</sup> hemi-Fontan or bidirectional cavopulmonary shunt, or both the latter procedures together<sup>9, 28, 29</sup> were not used, and variations of the total cavopulmonary connection or lateral atrial tunnel repair<sup>3, 5, 30</sup> were only used for anomalies of systemic or pulmonary venous drainage, as previously described.

**Outcome event.** The outcome event for this study was defined as takedown of the repair or death during the initial hospitalization or within 30 days of the operation.

**Statistical analysis.** Associations between the outcome event and 33 clinical and hemodynamic variables (Tables I and II) were analyzed univariately by the  $\chi^2$  test or Fisher's exact test, or both, for dichotomous variables and the Wilcoxon rank-sum test for continuous and ordinal variables. A stepwise multiple logistic discriminant analysis was used to identify factors jointly associated with the outcome event after the modified Fontan operation. Variables were retained in the multivariate analysis if their *p* values were less than 0.05. The odds ratios and corresponding 95% confidence intervals for each statistically significant variable were then calculated on the basis of the estimated coefficients obtained from the logistic model.

From the final logistic model, an estimated predicted probability of having the outcome event was obtained for each patient. The sensitivity of the model was defined as the proportion of patients who had the event who each had a model-predicted probability higher than a given cutoff value, whereas specificity was the proportion of patients who did not have the event who had model-predicted probabilities below the cutoff value.

## Results

The mortality rate during the initial hospitalization or within 30 days of the operation was 14.0% (*n* = 98) and an additional 6 of 16 patients survived takedown of the repair, for a combined event rate of 14.8% (*n* = 104). This rate has significantly dimin-

ished over the years from 26.6% before 1980 to 13.0% since 1980 (*p* < 0.001) (Table III) despite the increased number of high-risk patients currently undergoing operation.

**Univariate analysis.** Patients who had the outcome event were more likely to have a complex form of functional single ventricle, asplenia, ligation of the pulmonary artery, higher intraoperative right atrial pressure (before and after the Fontan operation) and higher intraoperative left atrial pressure (after the Fontan procedure), higher preoperative and intraoperative (before Fontan procedure) pulmonary artery mean pressure, longer aortic cross-clamp time, larger heart size (preoperative cardiothoracic ratio), higher ventricular end-diastolic pressure, and younger age and lower body weight at the time of operation (Tables I and II, *p* < 0.05). Concomitant repair or replacement of the atrioventricular valve was of marginal significance (*p* = 0.057).

A new variable, "corrected pulmonary artery pressure," was also examined. This is defined as mean pulmonary artery pressure divided by the ratio of pulmonary to systemic flow (Qp/Qs) (if Qp/Qs is greater than 1.0) or, if Qp/Qs is 1.0 or smaller, mean pulmonary artery pressure divided by unity. Although this was highly significant (*p* = 0.002), it was no more predictive than preoperative pulmonary artery mean pressure and therefore was not included in the multivariate model.

**Multivariate analysis.** On the basis of a stepwise logistic discriminant analysis, seven variables were identified as associated with the outcome event of interest: year of operation before 1980, presence of asplenia, younger age at operation, higher preoperative pulmonary artery mean pressure, longer aortic crossclamp time, pulmonary artery ligation, and higher intraoperative right atrial pressure (after Fontan procedure) (Table IV). At a specificity of 80%, the sensitivity for this model was 73%.

## Discussion

The Fontan-type repair may be the definitive palliation for a variety of complex cyanotic heart defects. During the past decade, the indications for the modified Fontan operation were extended to patients with more complex lesions<sup>2, 4-6, 15-16, 24-25</sup> and to patients who fulfilled fewer and fewer of the widely accepted criteria for low-risk operation.<sup>7, 17-19, 21, 31</sup> Associated with this enthusiasm for the Fontan repair came a better understanding of the hemodynamics and pathophysiologic features of the repair<sup>5, 26, 32-37</sup>

**Table I.** Univariate relations between dichotomous or ordinal demographic/clinical variables and early postoperative death or takedown

Factor (% missing)*	No. of patients	Takedown and/or hospital death	
		%	p Value
Gender			
Male	422	16	0.450
Female	280	14	
Lesion			
Complex	273	19	0.012
Other	429	12	
Polysplenia			
Yes	46	22	0.171
No	656	14	
Asplenia			
Yes	46	35	<0.001
No	656	13	
Enlargement of bulbar ventricular foramen			
Yes	59	17	0.630
No	643	15	
Left and/or right pulmonary artery reconstruction			
Yes	154	14	0.834
No	548	15	
AV valve repair or replacement			
Yes	66	23	0.057
No	636	14	
Preoperative AV valve insufficiency			
None	459	15	0.620
Mild	152	13	
Moderate	45	16	
Severe	46	22	
Preoperative pulmonary AV fistula			
Yes	18	17	0.740
No	684	14	
Division of pulmonary artery			
Yes	257	12	0.119
No	445	16	
Ligation of pulmonary artery			
Yes	245	20	0.005
No	454	12	
AV valve patch closed			
Yes	228	15	0.970
No	466	15	
Preoperative atrial arrhythmia			
Yes	66	12	0.518
No	636	15	
Valved connection			
Yes	52	13	0.773
No	650	15	

**Table I. Cont'd**

Factor (% missing)*	No. of patients	Takedown and/or hospital death	
		%	p Value
Pulmonary arteriolar resistance index (39%)			
<2	297	14	0.238
2-4	117	16	
5+	13	31	
NYHA functional class (32%)			
I	8	13	0.121
II	252	15	
III	207	20	
IV	9	22	
Intraoperative (after Fontan) RA pressure			
>20	110	37	<0.001
≤20	574	9	
Intraoperative (after Fontan) LA pressure			
>10	273	19	0.001
≤10	387	10	
Ventricular end-diastolic pressure (15%)			
>15	158	16	0.664
≤15	440	15	
Ejection fraction (52%)			
≤50	104	14	0.347
>50	231	11	
Age (yr)			
<4	84	25	0.005
≥4	618	13	
Weight (kg)			
≤15	131	24	0.001
>15	567	13	

All variables are preoperative measurements except for intraoperative right and left atrial pressure measurements. AV, Atrioventricular; NYHA, New York Heart Association; RA, right atrial; LA, left atrial.

\*Percentage of patients with missing data.

and improved perioperative management, all of which translated into better early results. As experience was gained, newer surgical innovations became available to deal with the most complex of lesions,<sup>2-5, 8-9, 15, 30, 38</sup> to reduce the frustrating early morbidity related to the postoperative accumulation of persistent or recurrent serous fluids,<sup>5, 10-11, 26, 39-40</sup> to improve the hemodynamics by possibly streamlining the systemic venous channels,<sup>5, 30</sup> and to support the patient during fluctuations in cardiac output and pulmonary vascular resistance.

More recently, we<sup>21, 23</sup> and others<sup>10, 41-42</sup> have cautioned against the application of the Fontan principle to patients who were less than ideal candidates for the operation, indicating that not only

**Table II.** Univariate relations between continuous demographic/clinical variables and early postoperative death or takedown

Factor	Overall percent missing	Early postoperative death or takedown						p Value
		No (n = 598)			Yes (n = 104)			
		Median	Mean	Range	Median	Mean	Range	
Age (yr)	0	9.0	11.0	0.66-42.0	6.0	8.0	0.66-29.0	<0.001
No. of prior procedures	0	1	1.3	0-6	1	1.5	0-7	0.181
Intraoperative (before Fontan) RA pressure (mm Hg)	31	8	8.5	1-25	9	9.9	2-23	0.046
Intraoperative (before Fontan) PA mean pressure (mm Hg)	47	13	14.2	3-53	16	18.1	5-58	0.002
Intraoperative (after Fontan) RA pressure (mm Hg)	3	18	17.7	8-30	20	20.2	9-34	<0.001
Intraoperative (after Fontan) LA pressure (mm Hg)	6	10	9.9	2-23	11	11.5	3-25	<0.001
Crossclamp time (min)	8	68	71.1	4-199	76	79.9	5-189	0.026
Preoperative CT ratio	10	0.55	0.55	0.29-0.81	0.59	0.58	0.42-0.82	0.003
Preoperative PA mean pressure (mm Hg)	15	16	18.0	2-73	19.5	22.2	8-58	<0.001
Preoperative Qp/Qs	33	1.18	1.56	0.15-9.00	1.34	1.57	0.22-6.50	0.921
Pulmonary arteriolar resistance index (mm Hg/L/min · m <sup>2</sup> )	39	1.4	1.6	0.1-5.6	1.5	1.8	0.2-4.5	0.247
Total pulmonary resistance (mm Hg/L/min · m <sup>2</sup> )	43	3.7	4.1	0.5-11.6	3.8	4.4	0.8-14.0	0.288
Ventricular end-diastolic pressure (mm Hg)	15	12	12.7	5-32	14	14.0	5-34	0.023
Preoperative ejection fraction (%)	52	57	55.7	30-82	55.5	56.0	34-78	0.852
Weight (kg)	1	26.2	31.8	5.9-99.0	18.5	24.0	5.7-79.5	<0.001
PA mean pressure/(Qp/Qs)*	37	12.0	12.6	0.9-34.8	13.0	14.9	2.4-30.4	0.002

RA, right atrial; PA, pulmonary artery; LA, left atrial; CT, cardiothoracic.

\*If Qp/Qs &gt;1 or pulmonary artery mean pressure/1; if Qp/Qs ≤1.

**Table III.** Distribution of early postoperative death or takedown by year of operation

Year of operation	No. of patients	Takedown and/or hospital death	
		No.	%
1973	1	0	—
1974	3	2	67
1975	4	1	25
1976	5	1	20
1977	10	2	20
1978	19	2	11
1979	52	17	33
Subtotal	94	25	27
1980	41	9	22
1981	48	9	19
1982	60	11	18
1983	50	5	10
1984	59	6	10
1985	70	9	13
1986	74	11	15
1987	66	8	12
1988	73	3	4
1989	67	8	12
Subtotal	608	79	13
Total	702	104	15

was the attrition rate high during the first 6 months after operation, but also that late complications developed in a significant proportion of patients after the repair, including arrhythmias,<sup>23-43</sup> protein-losing enteropathy,<sup>44-46</sup> and progressive cardiac failure and death.<sup>20, 23, 41-42</sup> Recent modifications such as the bidirectional Glenn anastomosis, fenestrated Fontan procedure, and adjustable atrial-septal defect after the Fontan operation may reduce significantly the perioperative morbidity and mortality. Whether this will translate into better long-term survival is unfortunately still unknown.

In the long term there may be advantages to earlier operations such as minimizing the deleterious effects of ventricular hypertrophy, chronic volume overload, and deteriorating cardiac function.<sup>17, 41, 42</sup> However, in our experience, which spans 16 years and in which two surgeons (G. K. D. and F. J. P.) each performed more than 300 modified Fontan operations on patients whose ages ranged from 8 months to 42 years, patients younger than 4 years still have a greater risk of dying or having the repair taken down after operation than do older patients. The discrepancy of this risk,

**Table IV.** Summary of logistic discriminant analysis

Variable	Estimated beta coefficient	Delta	Odds ratio* (95% CI)	p Value
Intraoperative (after Fontan) RA pressure	0.235	2	1.60 (1.31, 1.94)	<0.001
Operation before 1980 (yes vs. no)	1.649	1	5.20 (2.45, 11.04)	<0.001
Age (yr)	-0.096	-3	1.33 (1.14, 1.57)	<0.001
Preoperative PA mean pressure	0.060	3	1.20 (1.10, 1.30)	<0.001
Asplenia (yes vs. no)	1.080	1	2.94 (1.17, 7.40)	0.022
Crossclamp time (min)	0.011	8	1.09 (1.01, 1.18)	0.024
Ligation of PA (yes vs. no)	0.605	1	1.83 (1.04, 3.22)	0.036

CI, confidence interval; RA, right atrial; PA, pulmonary artery.

\*Estimated odds of having an event per delta unit change in the value of each variable.

however, has decreased with more recent year of operation. Most surgeons currently think that the optimal age for the modified Fontan operation in uncomplicated cases should be 2 to 4 years.<sup>7, 17-19, 47</sup> Our data may be influenced adversely by the disproportionate number of patients with many risk factors (in addition to younger age). Indeed, we have reported that in patients at low risk the mortality for the 2- to 4-year-old patients is no different from that of older patients.<sup>18</sup>

However, it is important to emphasize that a major limitation of a retrospective study such as this one is the unavailability of complete data on all patients. For example, a measurement of preoperative ejection fraction was unavailable for 52% of the patients in this study. In addition, a clinical selection process may exclude patients with presumed adverse risk factors from the cohort. Both of these factors will prevent the identification of important predictive variables.

Although higher pulmonary arteriolar resistance is unquestionably an important risk factor, we could not identify it as an independent variable in this analysis. This may reflect both the narrow range within which patients are accepted for operation and the high proportion ( $\pm 40\%$ ) of patients in whom data for this variable were missing. It is reasonable, however, to assume that both higher preoperative pulmonary artery mean pressure and higher postoperative right atrial pressure (both highly significant determinants of an event in this analysis) reflect elevated pulmonary vascular resistance.

We were somewhat surprised to find that the new variable corrected pulmonary artery pressure was not more predictive of an event than simply the use of the pulmonary artery mean pressure. Again, the 37% missing data may have diminished the statisti-

cal significance of this new concept. Motivation for the development of this variable derived from the clinical situation in which a patient with a pulmonary artery pressure of, for example, 24 mm Hg is evaluated for operation. If the Qp/Qs in this patient is, for example, 2:1 and this is restored to 1:1 by the operation, then the pulmonary artery pressure may be expected to be reduced to 12 mm Hg (24/2) after operation.

The need for reconstruction of the pulmonary arteries, significant systemic atrioventricular valve regurgitation, and the number of previous operations were not determinants of an event in this study. Because the follow-up period was restricted to hospital discharge or 30 days after operation, we did not assess whether these factors were still predictive of a poor long-term outcome.<sup>23</sup>

Though our results with this subgroup have improved over the years,<sup>4</sup> the heterotaxia asplenia syndrome remains a significant determinant of an event in this study. This reflects the complex cardiac pathologic features present and the complicated intracardiac baffles necessary to separate the circulations. However, the significance of pulmonary artery ligation as a risk factor is less clear: 502 of the 702 patients in this study had either ligation ( $n = 245$ ) or division ( $n = 257$ ) of the main pulmonary artery, and although division of the main pulmonary artery is not statistically important ( $p = 0.119$ ), ligation of the pulmonary artery was significant both univariately ( $p = 0.005$ ) and multivariately ( $p = 0.036$ ). Whether this is a confounding variable, a positive variable (for example, ligation may occasionally result in some distortion of either the branch pulmonary arteries or even closely related coronary arteries), or a negative variable (for example, *not* ligating the pulmonary artery and allowing a small amount of prograde flow through a severely

obstructive pulmonary artery may be beneficial in the short term) is extremely difficult to determine in a retrospective analysis such as this.

From the multivariate analysis it is evident that younger age, higher preoperative pulmonary artery mean pressure, higher postrepair right atrial pressure, early year of operation, presence of asplenia syndrome, longer aortic crossclamp time, and ligation of the pulmonary artery are important determinants of an event in this analysis (Table IV). For this model at a specificity of 80%, the sensitivity was 73%.

One should, however, be cautious when drawing conclusions from this study, because the end point for an event is early postoperative death or Fontan takedown. Although this allows for easy comparison with other studies, a good outcome by this definition does not necessarily reflect a satisfactory overall result, because the attrition rate is known to be higher for the first 6 months after operation.<sup>21, 41-42</sup> This is an important major limitation of an analysis such as this. However, the objective of the study was to relate the risk analysis for early postoperative death or takedown in our patient cohort to that of previous studies to determine whether any risk factors have been nullified by experience and time, both within our practice and relative to other large series.

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